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Case Report

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Familial Complete Atrioventricular Block (FCAVB) DDDR Pacemaker Implant with Synchromax

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ABSTRACT

We present the case of a family with complete atrioventricular block involving several generations, with autosomal dominant Mendelian inheritance, where one of those long-term affected presents heart failure. This entity corresponds to progressive familial heart block type I, mainly described in South Africa. Permanent Pacemaker implant using Synchromax was successful.

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Introduction

In Venezuela to date, we are not aware of published cases of familial complete atrioventricular block (FCAVB) and the use of the Synchromax device as a method to prevent pacemaker-induced cardiomyopathy.

The literature on the origin of complete AV block (CAVB) in young people is scarce, only between 3-5% of all patients undergoing pacemaker implantation are among 18-55 years of age [1].

The first case of FCAVB was published by Morquio in 1901. Since then, numerous studies on FCAVB have been published and several genes implicated in this familial heart disease have been identified [2,3].

We present the case of a 30-year-old patient who was admitted to our department with syncope due to CAVB, and during the interview we found family history of CAVB.

Case Report

The index case corresponds to a 30-year-old male patient who was admitted to our service due to syncope with CAVB, who also referred to the interview of a decrease in his functional capacity for 1 year. Relevant family history includes 56-year-old father with a DDDR permanent pacemaker implant since the age of 45 due to CAVB and a 75-year-old living maternal grandmother with a DDDR permanent pacemaker since she was 54 years old. (Figure 1). The electrocardiogram (ECG) of the index case showed a CAVB rhythm with a wide QRS ventricular escape (Figure 2), the ECGs of the father and the paternal grandmother showed a DDDR pacemaker rhythm compatible with the ventricular electrode positioned at the apex of the right ventricle (figure 3). The echocardiograms of the index case and his father were normal. Paternal grandmother´s echocardiogram showed moderately depressed ejection fraction (45%) and a 45mm slightly dilated left atrium. Chagas disease was ruled out in all 3 patients.

In the index case, an ENDURITY DR ABBOTT pacemaker was implanted using the Synchromax device as a guide to position the ventricular lead in a para-Hisian zone with a synchrony curve of less than 0.40. Father and grandmother showed ventricular asynchrony curve when studied with Synchromax.

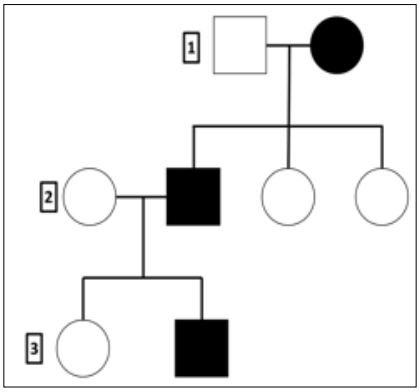

Figure 1: Square Male. Circle Female. Level 1 Grandmother. Level 2 Father. Level 3 Index Case

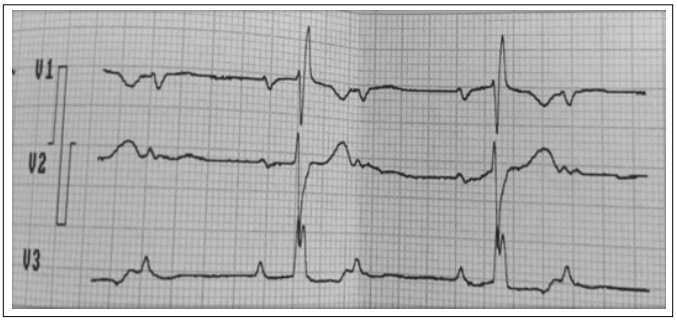

Figure 2: FCAVB simulating AVB 2:1 with Right Bundle Branch block

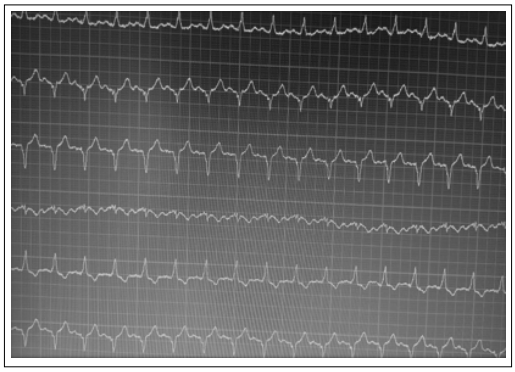

Figure 3: DR pacing rhythm stimulates left ventricular apex

Discussion

In the Framighan study, a high-risk ratio for pacemaker implantation could be observed in patients with first-degree relatives who developed atrioventricular blocks and bundle branch blocks, suggesting a hereditary nature of the total atrioventricular block [4], in this instance, the index case at the age of 30 was admitted due to syncope and had a paternal family history of CAVB. In this same study, it was found that the incidence of pacemaker implantation in people under 50 years of age is 17.7/year/million inhabitants in a follow-up period of 20 years. The etiology of CAVB was identified in less than half of the patients also suggesting that in many cases the cause of the implant is unknown because genetic tests are not performed. Genetic tests could be performed in all patients under 50 years of age who undergo a pacemaker implantation when the cause is unknown. Unfortunately, in our country, routine genetic tests are not yet performed in the recognition of channelopathies and the diagnosis is based on the patient's medical history [3].

Resdal Dideriksen et al. report that in the long-term follow-up patients with an implantation age of less than 50 years due to CAVB of unknown origin have 4 times greater risk of presenting heart failure, death, and hospitalizations than the control group, and this risk begins to manifest 5 years after the implant [5].

In the neighboring country of Colombia, Mora et al report the case of a family with atrioventricular block involving several generations with autosomal Mendelian inheritance, which progressively affects the cardiac conduction system leading to syncope at an early age. This entity corresponds to a type I FCAVB, described mainly in South Africa. Our case is representative of this pathology since it is a young adult with a rich family history and a wide QRS FCAVB. Type I FCAVB is characterized by the fact that the escape rhythm exhibits a wide QRS and type II displays a narrow QRS [6].

Conventional cardiac pacing in the apex of the right ventricle effectively restores the patient's heart rate; however, it causes serious long-term electromechanical problems due to a stimulated QRS with left branch morphology, which induces dyssynchromyopathy due to induction of mitral regurgitation, adverse cardiac remodeling and ventricular systolic dysfunction [7] as in the case of the index patient's grandmother who presented a moderately depressed systolic ejection fraction.

To prevent the development of heart failure in the long term we used the para-Hisian pacing technique with the aid of the Synchromax Exxo Argentina device.

The conventional bipolar ventricular electrode was taken to the outflow tract of the right ventricle into the pulmonary artery and then slowly retracted and positioned in an area where the equipment showed an electrical synchrony curve of less than 0.4 with excellent electronic parameters. This equipment performs a computerized graphic and mathematical processing of averaged signals taking as references the leads DII (right interventricular septum) and V6 (left ventricular lateral wall) the result of these mathematical calculations generates 3 types of curves, values less than 0.40 are considered physiological, from 0.41 to 0.69 mild dyssynchrony and greater than 0.7 severe dyssynchrony [8].

Currently, in the three-month follow-up, the patient remains clinically stable, maintaining a physiological electrical synchrony curve and preserved ejection fraction. We hope to evaluate his electromechanical evolution in the long term, since, as previously described, the main problems occur after 5 years of the implant [5].

Conclusion

This is the first case reported in Venezuela of FCAVB 1; the interview is the main tool we have in our country to make this diagnosis since by not having a genetic test available we must highlight the importance of the patient's clinical history. We will evaluate in the long term the usefulness of Synchromax in the prevention of dyssynchrony.

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